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Case Report

Hemiagenesis of unilateral lobe of thyroid gland

Sameer Karmacharya [1], Santosh Kumar Sah[1], Sunil Adhikari[2]

Department of Otorhinolaryngology and Head and neck surgery
Janaki Medical College Teaching Hospital
Ramdaiya, Janakpur

[1]Assistant Professor, Department of Otorhinolaryngology and Head and neck surgery, JMCTH
[2]Assistant Professor, Department of Radiodiagnosis, JMCTH

ABSTRACT

The thyroid gland begins its development during the 3rd week of gestation from the median endodermal thickening in the floor of the pharynx that later form the median diverticulum. During development, the thyroid gland normally descends to its typical location in front of the pharynx. Failure of descent has been suggested as a cause of thyroid hemiagenesis; however, the compensatory growth of the other lobe suggests a lobulation defect rather than a failure of descent. Abnormalities during the development lead to defective organogenesis or descent, complete or partial absence of the gland with or without ectopic thyroid tissue. Thyroid hemiagenesis is a rare congenital abnormality characterized by the absence or failure to develop one lobe of the thyroid gland with or without the absence of the isthmus. Unilateral thyroid agenesis (thyroid hemiagenesis) is a rare disorder with less than 300 cases currently in the world literature. Here, we report a case of left hemiagenesis of thyroid.

Key words: Thyroid gland, Hemiagenesis, congenital abnormality, Lobulation defect

INTRODUCTION

During development, the thyroid gland normally descends to its typical location in front of the pharynx [1]. Failure of descent has been suggested as a cause of thyroid hemiagenesis; however, the compensatory growth of the other lobe suggests a lobulation defect rather than a failure of descent [2]. Thyroid hemiagenesis has been reported to occur in 0.05-0.2% of the population with Sakura et al noting in 2007 the incidence of 250 reported cases. It occurs more commonly in females [3]. Thyroid agenesis is often an incidental finding when the thyroid gland is being imaged for suspected pathology or during a screening examination. If no thyroid pathology is demonstrated in the existing lobe, one lobe of the thyroid is adequate to maintain clinical euthyroidism [4].

Case Report

A 13 year old female presented with right sided neck swelling noticed by her father 21 days back. It was painless and was not associated with any compressive symptoms like dyspnea, dysphonia, or dysphagia. There was no history of tremors, palpitations, or change in weight or bowel habits . No family history of hypothyroidism or hyperthyroidism.

Clinical examination of the neck revealed a soft swelling in the region of the right lobe of thyroid which moved freely with deglutition (Figure 1). There was no cervical

lymphadenopathy and no eye or skin changes. Systemic examination was normal.

Investigations were as follows:

ESR < 15mm/hr

Thyroid function test

Free T4 - 10.2 pg/ml (normal 8.9 – 17.2),

Free T3- 2.8 pg/ml (normal 1.21- 4.18),

TSH – 3.2 µIU/ml (normal 0.35- 4.35).

Figure 1: Compensatory enlargement of the right lobe in a patient with left-sided thyroid hemiagenesis

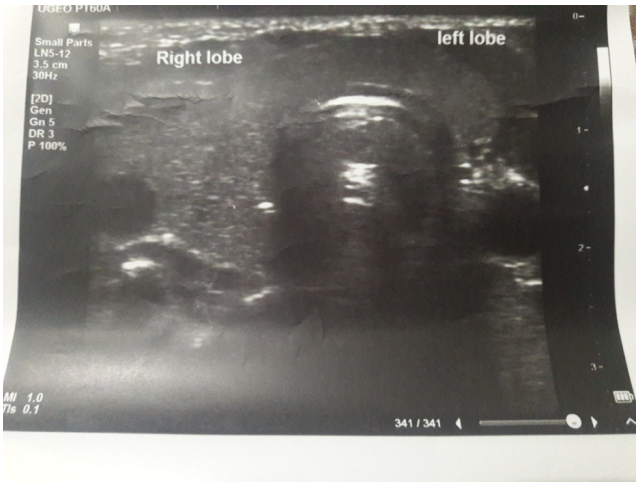

Figure 2: Ultrasound picture of left-sided thyroid hemiagenesis in a transverse section of the neck. Right lobe and isthmus are present

Ultrasound neck showed enlarged isthmus and right lobe of thyroid gland with normal outline, ehotexture and vasularity. Left lobe of thyroid gland was hypoplastic suggestive of hemiagenesis of left lobe of thyroid gland with hypertrophied isthmus and right lobe of thyroid gland (Figure 2).

DISCUSSION

Developmental anomalies of the thyroid gland are rare, and are usually due to the result of abnormal descent of the thyroid gland rather than abnormal development. Hemiagenesis is a form of thyroid dysgenesis, in which one thyroid lobe fails to develop, with or without agenesis of the isthmus. It was first described in 1866 by Handsfield-Jones[5]. The absence of one lobe of the thyroid commonly has no clinical significance or symptoms . However, reports have suggested that in adults with hemiagenesis, the development of compensatory hypertrophy of the contra- lateral thyroid tissue, thyroid nodular disease, disturbances in thyroid function, thyroid morphological abnormalities, and thyroid autoimmune disorders are more likely to develop. The prevalence of these findings increases significantly with age[6]. Nodules discovered inadvertently during an unrelated examination are defined as thyroid incidentalomas. Sonographic examinations have described detection of incidentalomas in 67% of patients [7]. Two studies involving 494 and267 participants, reported malignancy rates of nonpalpable thyroid nodules at 9% and 12%, respectively . Management of thyroid nodules is controversial . It is recommended that nodules with a diameter <1 cm and without a suspicious history or ultra- sound findings should have a follow-up ultrasound examination and thyroid stimulating hormone (TSH) measurements in 6-18

months[8]. Sonography is the most cost-effective means of diagnosing thyroid hemiagenesis[9]. Computed tomography (CT) and magnetic resonance imaging (MRI) may also be used, but are expensive and time-consuming in comparison. Thyroid hemiagenesis is commonly diagnosed on thyroid scintigraphy with the accumulation of tracer in only one lobe of the thyroid [10].

Thyroid scintigraphy is the only imaging modality that assesses thyroid regional function while detecting areas of autonomously functioning thyroid tissue.

Clinicians should be aware of this developmental anomaly as hypothyroidism can be associated with this condition and also to prevent inadvertent surgical removal of the enlarged functioning lobe of the gland. Early screening and treatment can prevent the complications of untreated hypothyroidism.

CONCLUSION

Thyroid hemiagenesis is a rare entity that is readily detected by sonography. Typically the patient is asymptomatic and thyroid function is normal. Detecting and understanding the clinical and imaging presentation of thyroid hemiagenesis may ensure timely diagnosis and appropriate management. Based on current literature, there is a real possibility that these patients may be at an increased risk for malignancy and/or functional disturbances in the remaining lobe. This important clinical question can only be answered by a properly powered study which will need a large population of screened cases with a proper duration of follow-up.

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Corresponding author:
Dr Sameer Karmacharya
Department of Otorhinolaryngology and Head and neck surgery
Janaki Medical College Teaching Hospital
Ramdaiya , Janakpur, Nepal.
Email: karmacharyasameer1234@gmail.com